We claim:

1. A method for treating a polyglutamine disease, comprising administering a compound selected from the group consisting of L-methionine S-sulfoximine, L-ethionine S-sulfoximine, glufosinate and branched chain α-keto acids derived from leucine, isoleucine or valine, to a patient in need of such treatment.

2. The method according to claim 1, wherein said polyglutamine disease is selected from the group consisting of Huntington's disease, spinocerebellar ataxia, and spinobulbar muscular atrophy.

3. The method according to claim 1, wherein said compound is L-methionine S-sulfoximine or L-ethionine S-sulfoximine administered orally, intravenously, or intrathecally.

4. The method according to claim 1, wherein said L-methionine S-sulfoximine or L-ethionine S-sulfoximine is administered intrathecally at a dosage between 1.0-5.0 mg/kg per 6-10 days.

5. The method according to claim 1, wherein said L-methionine S-sulfoximine or L-ethionine S-sulfoximine is administered orally or intravenously at a dose between 2.0-10.0 mg/kg per 6-10 days.

6. The method according to claim 1, wherein said compound is glufosinate administered intrathecally at a dose of 1.0 – 5.0 mg per 6 -10 days.

7. The method according to claim 1, wherein said compound is an α-keto acid derived from leucine, isoleucine or valine.

8. The method according to claim 7, wherein said α-keto acid is selected from the group consisting of α-keto-isocaproate, α-keto-β-methylbutyrate and α-keto-valerate and salts thereof.

9. The method according to claim 7, wherein said α-keto acid is administered in a dosage between 280-380 mg/kg body weight.

10. The method according to claim 1, further comprising administering a second compound which inhibits aggregate formation, inhibits transglutaminase, inhibits caspase, or is neuroprotective.

11. The method according to claim 10, wherein said second compound is selected from the group consisting of Congo red, cystamine, cysteamine, minocycline, ethyl eicosapentaenoate, and riluzole.

12. A composition comprising a) an amount of a compound, selected from the group consisting of L-methionine S-sulfoximine, L-ethionine S-sulfoximine,

glufosinate and branched chain α-keto acids derived from leucine, isoleucine or valine, effective to treat a polyglutamine disease, b) a second neuroprotective compound, and c) a pharmaceutically acceptable carrier.

13. The composition according to claim 12, wherein said second neuroprotective compound inhibits: aggregate formation, transglutaminase and/or caspase.

14. The composition according to claim 12 wherein said second compound is selected from the group consisting of Congo red, cystamine, cysteamine, minocycline, ethyl eicosapentaenoate, riluzole, L-methionine S-sulfoximine, L-ethionine S-sulfoximine, glufosinate and branched chain α-keto acids derived from leucine, isoleucine or valine.

15. A kit comprising two or more compounds selected from the group consisting of L-methionine S-sulfoximine, L-ethionine S-sulfoximine, glufosinate and branched chain α-keto acids derived from leucine, isoleucine or valine, in separate containers.

16. The kit according to claim 15, further comprising another compound useful for the treatment of polyglutamine diseases.

17. The kit according to claim 16, wherein said compound useful for the treatment of polyglutamine diseases inhibits aggregate formation, inhibits transglutaminase, inhibits caspase, or is neuroprotective.

18. The kit according to claim 17, wherein said compound useful for the treatment of polyglutamine diseases is selected from the group consisting of Congo red, cystamine, cysteamine, minocycline, ethyl eicosapentaenoate, and riluzole.

19. A method for decreasing neuronal polyglutamine containing aggregates, comprising administering at least one compound selected from the group consisting of L-methionine S-sulfoximine, L-ethionine S-sulfoximine, glufosinate and branched chain α-keto acids derived from leucine, isoleucine or valine, to a patient in need of such decrease.

20. A method for decreasing the amount of huntingtin protein in brain tissue, comprising administering at least one compound selected from the group consisting of L-methionine S-sulfoximine, L-ethionine S-sulfoximine, glufosinate and branched chain α-keto acids derived from leucine, isoleucine or valine, to a patient in need of such decrease.